Inflammatory bowel disease and the craniocervical junction

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Rheumatological complications have been described in up to 30% of patients being followed for inflammatory bowel disease. The majority of these complications occur as spondylitic changes in the lumbar spine. Erosive arthritic disease associated with inflammatory bowel disease occurs less frequently, but it can result in ligamentous laxity and joint instability. To highlight the potential significance of the process at the craniocervical junction, the authors describe the long-term follow-up care of a complicated case. A 56-year-old woman, with a long history of ulcerative colitis, presented with atlantoaxial instability and underwent a C1-3 fusion; however, the presence of significant occipitoatlantal instability was not recognized. This resulted in high cervicomedullary quadriplegia, requiring traction reduction and a combined anterior transoral decompressive-posterior occipitocervical fusion. The patient's neurological deficit completely resolved postoperatively.

Key Words * inflammatory bowel disease * craniocervical junction * cervical spine * spondyloarthropathy * enthesopathy

The incidence of rheumatological involvement associated with inflammatory bowel disease varies from 2 to 30%, most often resulting in joint effusion or spondylitic changes in the lumbar spine.[1-7, 9,16,18] Inflammatory bowel disease in cases that involve arthritis is generally classified with the seronegative spondylarthritides, which also include psoriasis and Reiter's syndrome.[3,7,9,16] Because, in these conditions, the site of joint involvement is often where the ligament or tendon is inserted into the bone (enthesis), this group is also referred to as the enthesopathies.[4]

Whereas the enthesopathies typically result in stiffening or fusion of the involved joints (spondylitis), the associated arthritis can cause severe erosive changes in the ligaments and associated joints.[4,6] When this occurs in the craniocervical region, significant instability can result.[6,18] The association of these conditions with craniocervical instability cannot be overlooked, because early diagnosis is crucial to avoid neurological compromise. This instability can be present at both the atlantoaxial and occipitocervical interfaces, as has been clearly described in cases of rheumatoid arthritis.[14] When either form of instability goes unrecognized, failure to stabilize the abnormal segment as well as further neurological compromise can result.[8,10-15]

As an example, we review the case of a 56-year-old woman with ulcerative colitis who presented with
cervical pain and a mild quadraparesis and in whom the posterior C1-3 fusion procedure, which was performed to treat presumed isolated atlantoaxial instability, caused disastrous results. The results of this case highlight the necessity to recognize two important issues: 1) the association of inflammatory bowel disease and related conditions with potential involvement of the craniocervical junction and 2) significant occiptiocervical instability in association with atlantoaxial instability.

**CASE REPORT**

This 56-year-old woman with a long history of ulcerative colitis presented locally with progressive myelopathy. Magnetic resonance imaging revealed craniocervical junction (CVJ) compression and subluxation (Fig. 1) and the patient underwent a posterior C1-3 fusion procedure at another institution, in which a rib and wire construct were used while she was in a sitting position (Fig. 2). Postoperatively she was quadriparetic and required a ventilator to breathe.

![Fig. 1. Preoperative T1-weighted sagittal magnetic resonance image obtained at the CVJ. Note the significant anterior luxation of the occiput and significant anterior cervicomедullary junction compression.](image-url)
She was transferred to the University of Iowa for further evaluation. Upon arrival she was dependent on a
ventilator, with absent gag reflex, and experienced severe quadriplegia. A magnetic resonance image was
obtained that revealed significant anterior cervicomedullary compression with anterior dislocation of the
occipitoatlantal interface (Fig. 3).

Fig. 3. Postoperative T1-weighted sagittal magnetic resonance image obtained at the CVJ
following posterior C1-3 fusion. The area of anterior compression appeared to have
worsened, and this correlated with the patient's neurological deterioration.
After being placed in traction the patient's spinal alignment and neurological status improved. However, because her symptoms were not completely resolved with traction alone, we chose to perform an anterior transpalatal transpharyngeal decompressive procedure at the cervicomedullary junction; resection of the C-1 arch and the odontoid process was followed by occipitocervical fusion in which a contoured titanium loop and rib graft was placed (Fig. 4). Her neurological status improved dramatically during the 1st postoperative week, and she was immobilized in a halo vest for 3 months. At 5-year follow-up evaluation she was neurologically intact.

Fig. 4. Anteroposterior (left) and lateral (right) radiographs obtained at the CVJ following traction reduction, anterior decompressive surgery, and posterior stabilization, showing realignment of the occipitocervical junction.

DISCUSSION

The most common rheumatological complications of inflammatory bowel disease are inflammatory effusions of the large joints and reactive spondylitis.[1] The former complication is generally nondestructive and self limited, whereas the latter commonly results in spondylitic changes in the spine and sacroiliac joints.[5,9,16] The term "seronegative spondylarthritides" refers to those conditions that cause inflammatory conditions of the spine and sacroiliac joints, with an absence of rheumatoid factor.[3,7] These conditions include: ankylosing spondylitis, Reiter's syndrome, some forms of juvenile rheumatoid arthritis, some cases of psoriatic arthritis, some cases of postinfectious arthritis, and the form of arthritis associated with inflammatory bowel disease. In these conditions the anatomical site of involvement is the enthesis, which is defined as the ligamentous or tendonous insertion into bone.[4,6] This metabolically active transition region between two collagenous surfaces is particularly susceptible to torsion-related injury and can be the site of chronic inflammation. The conditions that result from inflammation of the entheses are referred to as enthesopathies, and they overlap with those of seronegative spondylarthropathies. In contrast, the central structure involved in rheumatic conditions is the synovial membrane.[14] Although synovial involvement can occur in the seronegative arthropathies, it is generally less severe and is believed to be secondary to the primary area of injury. Atlantoaxial instability, although common in cases of rheumatoid arthritis, is generally believed to be uncommon in the seronegative spondylarthritides.[6,18] For example, ankylosing spondylitis frequently affects the cervical spine in terms of fusion between vertebral bodies, but atlantoaxial luxation is less frequent. This relationship may be altered when ankylosing spondylitis is present in conjunction with a peripheral
disease such as psoriasis, Reiter's syndrome, or inflammatory bowel disease.[18]

The relationship between inflammatory bowel disease and craniocervical disease has not been frequently addressed in the medical literature. In 1986 Jordan, et al.,[6] claimed to report the first description of a case in which inflammatory bowel disease was associated with atlantoaxial instability. In this case a 21-year-old African-American woman presented with C1-2 instability and was ultimately diagnosed as having Crohn's disease. She had not sustained any neurological deficit and underwent successful halo vest therapy. These authors were the first to indicate that in addition to spondylitis, which is relatively common in patients with seronegative arthritides, inflammatory bowel disease can cause an erosive arthritis similar to the effects of rheumatoid arthritis. They concluded that inflammatory bowel disease should be added to the differential diagnosis of patients who present with isolated atlantoaxial instability.

However, in 1969, Newman and Sweetnam,[15] in describing their technique for occipitocervical fusion, had previously reported the case of a 25-year-old woman in whom ulcerative colitis and an inflammatory polyradiculopathy were diagnosed. She presented with severe neck pain and subluxation of the atlantoaxial joint. She underwent occipitocervical fusion, which resolved her pain. These authors underscored the importance of carefully considering incorporation of the occiput in cases in which significant upward displacement of the dens was present. The concept of cranial settling has subsequently been well characterized and is of particular relevance in the aforementioned case.[14,17]

In reviewing their series of patients with ankylosing spondylitis, Suarez-Almazor and Russell[18] observed a substantial increase in atlantoaxial instability in those patients with ankylosing spondylitis if it was associated with psoriasis, Reiter's syndrome, or inflammatory bowel disease. Of their 39 patients, in nearly one-third of those with other peripheral disease atlantoaxial instability was demonstrated. In addition to spondylitic changes, these authors also noted the presence of an erosive form of arthritis that caused instability. Interestingly, in one of their patients, isolated erosion of the odontoid process was noted in the absence of atlantoaxial instability. They did not comment on the motion observed at the occipitoatlantal interface, but one could speculate that this case may have represented isolated occipitoatlantal instability without atlantoaxial instability, which has been described in other conditions associated with craniocervical anomalies such as Down's syndrome.[10]

In any patient with suspected atlantoaxial instability, careful assessment of potential occipitoatlantal instability and the extent of anterior compression is critical. The approach to diagnosing and treating patients with CVJ anomalies has been addressed comprehensively.[8,11-14] It is reasonable to initiate traction therapy in patients with craniocervical instability in the setting of a presumed inflammatory arthritis, because in the majority of cases at least partial decompression is possible. If satisfactory reduction can be confirmed both clinically and radiographically, a posterior fusion procedure may be sufficient to attain stability. Whether to incorporate the occiput remains the subject of some debate, but the decision is based on an assessment of the presence and degree of occipitoatlantal instability as well as the risk of developing cranial settling following C1-2 fusion alone.[8,11,14] If a significant component of anterior compression cannot be reduced, an anterior cervicomedullary decompressive procedure must be considered. In this setting an occipitocervical fusion is mandatory. As the results of our case illustrate, remarkable improvement and excellent long-term results can be obtained, even in the face of severe neurologic deficit, if the appropriate management principles are followed.

**CONCLUSIONS**

Inflammatory bowel disease can be associated with severe erosive arthritis that affects the CVJ. This
process, although infrequent, can result in severe neurological deficit if it is not recognized. The possibility of occipitoatlantal involvement in any inflammatory atlantoaxial subluxation must be considered in the selection of treatment strategies. The same principles applied to occipitocervical disease in cases of rheumatoid arthritis appear to be appropriate to guide management decisions in these unusual cases.

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